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CASE REPORT
Congenital heart disease

Percutaneous closure of an aortopulmonary window in a young adult patient: a case report of transcatheter closure with an occluder device

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Background Aortopulmonary window is a congenital heart disease requiring a surgical approach to achieve a complete correction.

Case summary We report on a case of a young female symptomatic adult patient with a significant aortopulmonary window. The patients refused surgical approach and since the defect was far enough from the pulmonary and aortic valve a percutaneous approach was proposed. The large aortopulmonary window was closed effectively with an Amplatzer Muscular Septal Occluder device (Abbott, Plymouth, MN, USA). At 1-year follow-up, the device was in a good position with no residual shunt.

Discussion Transcatheter closure of an aortopulmonary window is an effective and safe procedure in specific anatomic defects.

Keywords Aortopulmonary window • Septal occluder device • Interventional cardiology • Case report

Learning points

- Transcatheter closure of an aortopulmonary window is a feasible choice in specific anatomic subsets.
- Several occluder devices are designed to achieve an effective closure according to the anatomy of the defect.

Introduction

Aortopulmonary window is a congenital communication connecting the ascending aorta to the pulmonary artery. Surgery is the first choice to achieve the closure of the window; however, percutaneous treatment is very rarely a suitable strategy. Several reports describe a transcatheter closure of small aortopulmonary windows by using either devices designed for arterial duct closure such as Amplatzer Duct Occluder Type I, Type II, and Type II Additional Size (Abbott, Plymouth, MN, USA) or devices studied for vascular embolization such as Amplatzer Vascular Plug family (Abbott).[1–4] There are few cases reports about percutaneous closure of large aortopulmonary window in which greater devices were required.[5–9] In our case, a transcatheter approach was chosen to close a non-restrictive large aortopulmonary window using an Amplatzer Muscular Septal Occluder (Abbott).

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Timeline

Day 1	The patient was admitted to our institution for heart failure
Day 2	Echocardiographic diagnosis of aortopulmonary window was performed
Day 30	Transcatheter closure of aortopulmonary window was executed with an Amplatzer Muscular Septal Occluder device
Day 36	Discharge in good clinical condition, 6 days after the procedure

Case presentation

We report a case of a young female patient (25 years old and 52 kg) who came to our department since she developed dyspnoea on mild effort during pregnancy. The patient did not show a significant medical history. A mesocardial continuous murmur was appreciated, with a high differential pressure (systemic arterial pressure: 120/50 mmHg). Echocardiography was performed immediately and showed severe left ventricle dilatation (end-diastolic diameter 69 mm) with mild reduction of systolic function (left ventricle ejection fraction 45%). At parasternal short-axis view, a distal aortopulmonary window with significant left-to-right continuous shunt was detected (*Figure 1*). Systolic pulmonary artery pressure was 40 mmHg. Anti-congestive medical therapy (diuretics and angiotensin-converting enzyme inhibitors) improved clinical condition of the patient. The Heart Team discussed the case addressing the management to a surgical closure of aortopulmonary window. However, the patient refused the surgery, and so an interventional approach was proposed to close the defect, since the aortopulmonary window was far enough from the aortic and pulmonary valves. A detailed informed consent was obtained by the patient. After delivery, the cardiac catheterization was performed.

The aortography showed an aortopulmonary window with significant and early opacification of pulmonary artery. The large defect (basal measure 9 mm) was far away from the aortic and pulmonary valve allowing a percutaneous chance to close without interference with ventricular-arterial valves function. A normal pulmonary pressure (mean pulmonary arterial pressure 20 mmHg) and pulmonary vascular resistance (1.10 WU) were recorded. A haemodynamically significant left-to-right shunt (Qp:Qs 2.10) was detected with an indirect Fick method. From arterial side, the window was crossed with a hydrophilic Terumo wire, which was captured by an Amplatz Goose Neck snare (Microvena Corporation, White Bear Lake, MN, USA) and pulled up to the femoral vein (outside the venous sheath) creating an arterial-venous circuit. From femoral vein, a sizing balloon was carried until the defect and inflated; highlighting the presence of significant waist measured 12 mm. The stretching confirmed the low distension of window and the percutaneous closure feasibility. An Amplatzer Muscular Septal Occluder device 16 mm (Abbott) was chosen. The unpredictability of device deployment suggested to use two safety measures: the over-the-wire technique and the capturing of device distal-end with an Amplatz Goose Neck snare (Microvena Corporation), as previously described.[10,11]

These measures allowed a hooking of device even after the realizing so that it might be retired in case of embolization. The device was deployed, and stability was checked with push-and-pull manoeuvre and then the delivery cable was unscrewed. No changes of device placement were noted and so both the snare and the wire were removed. Both aortography as well as pulmonary arteriography confirmed a good position of the device without residual shunt or injuries on great vessels or coronary arteries (*Figure 2*).

The patient was extubated and discharged 1 day and 6 days later, respectively. No-inotropic support was required during hospitalization.

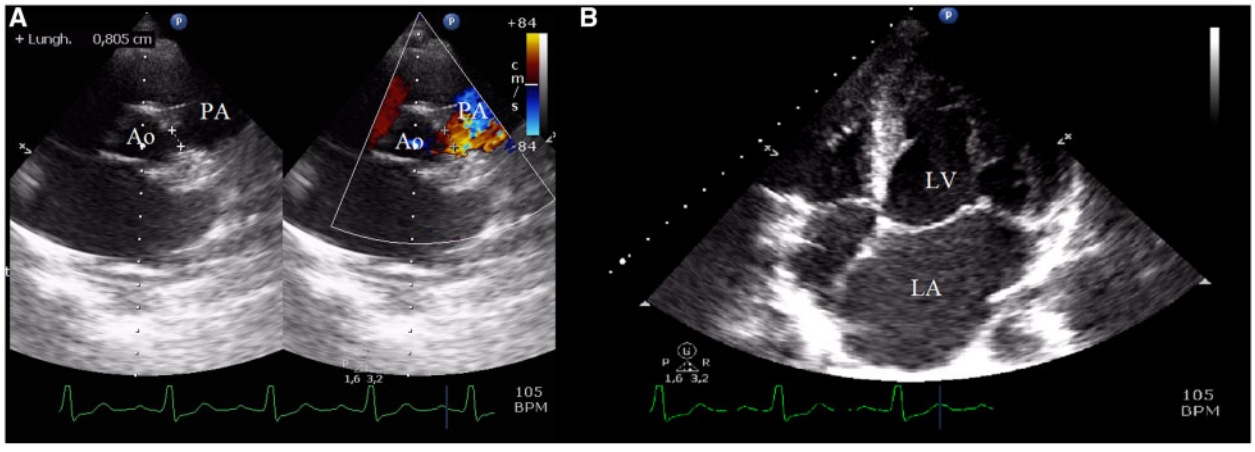

Figure 1 Pre-operative transthoracic echocardiography. (*A*) Parasternal short-axis view showed a large aortopulmonary window (~8 mm) with moderate left-to-right shunt from the aorta to pulmonary artery. (*B*) Apical four-chamber view demonstrated a severe dilation both of left atrium as well as of left ventricle. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery.

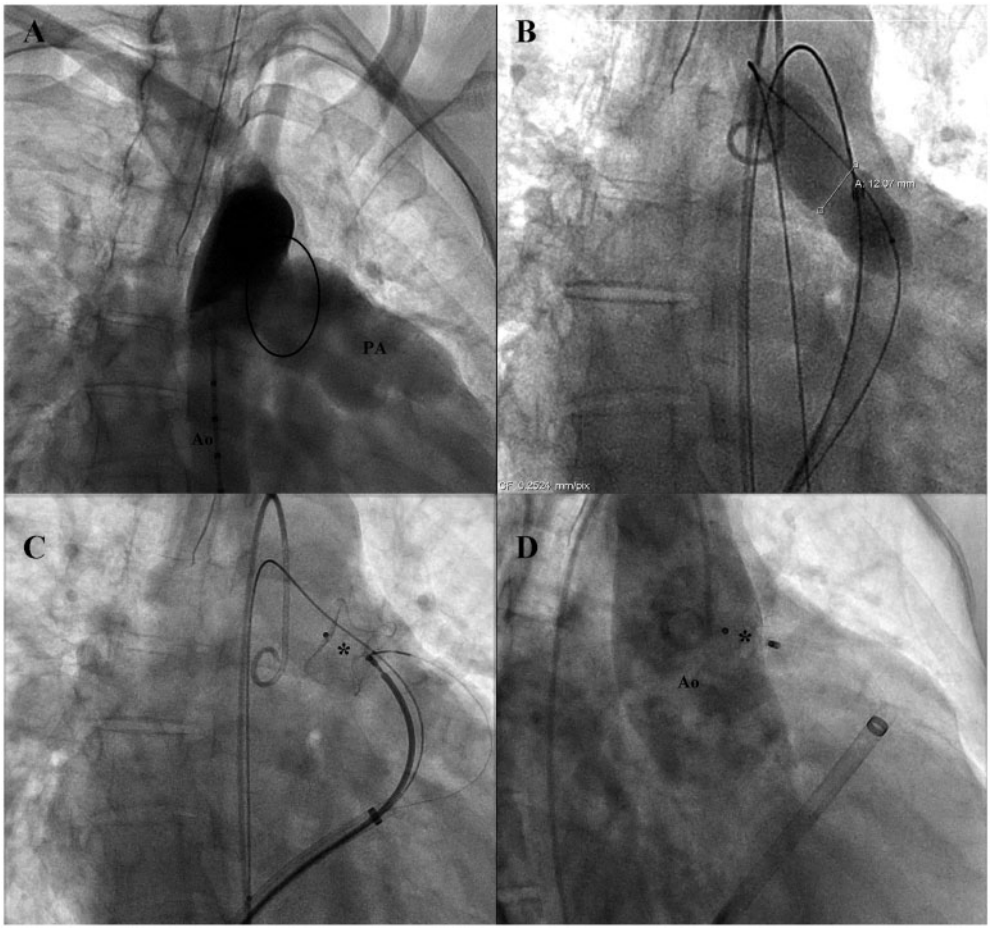

Figure 2 Transcatheter closure of the aortopulmonary window. (*A*) The aortic angiography (right anterior oblique view) showed a large aortopulmonary window (circled) with significant left-to-right shunt between the aorta and the pulmonary artery. (*B*) The aortopulmonary window was stretched with a sizing balloon with a waist measured 12 mm. (*C*) An over-the-wire technique and the capturing of device distal-end with an Amplatz Goose Neck snare were strategies used to deploy an Amplatzer Muscular Septal Occluder device 16 mm (*) (Abbott, Plymouth, MN, USA). (*D*) After device realizing, the final angiography demonstrated a complete closure of aortopulmonary window without residual shunt. Ao, aorta; PA, pulmonary artery.

At last follow-up (1 year later), the patient was asymptomatic, and echocardiography confirmed the good deployment of device with the absence of peri-prosthesis shunt (Figure 3) and normalization both of left ventricle diameter as well as of ejection fraction. Normal pulmonary pressures (systolic pulmonary arterial pressure 30 mmHg) were estimated through the tricuspid regurgitation.

Discussion

Aortopulmonary window is a rare congenital heart disease classified into three different types: Type I (proximal), Type II (distal), and Type III that is a large defect involving both proximally and distally the ascending aorta. Surgical repair is the primary treatment, particularly in newborns and infants. Percutaneous treatment is a less common choice. Devices designed for arterial duct closure or vascular embolization are ideal to achieve a closure of small aortopulmonary window and several reports describe it,[1–4] however, they are not useful to close major defects. In these last cases, atrial or ventricular septal occluder devices are required, however, proximal aortopulmonary windows may not be closed with these devices since these kinds of lesions are too much near to the cuspids of aortic and pulmonary valves and they might interfere with valvular function. Distal aortopulmonary window may be suitable of transcatheter correction. An adequate planning of procedure is needed. A balloon sizing of the window is required to stretch the defect and then to address the choice of the device. In our case, it was decided to overestimate the defect to ensure a perfect anchoring of the central waist of the device. Amplatzer Muscular Septal Occluder device 16 mm (Abbott) was chosen rather than an Amplatzer Septal Occluder device (Abbott) because characterized by less prominent disks. The less predictable final deployment of the device after the realizing required to adopt safety measures to avoid embolization. The over-the-wire technique and a preventive anchor of the device with a snare allowed

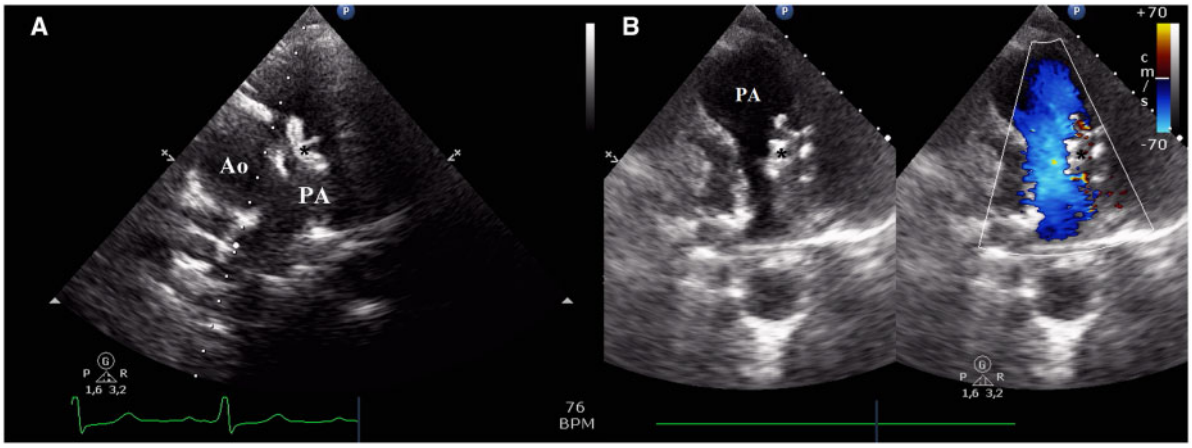

Figure 3 At last follow-up, the transthoracic echocardiography showed a good deployment of the device (*) without peri-prosthetic residual shunt from aorta to pulmonary artery and interference with aortic and pulmonary blood flow both in parasternal short-axis view (*A*) and in suprasternal view (*B*). Ao, aorta; PA, pulmonary artery.

both to have a complete control and to pull-back the device even after realized. In literature, this is the second case of aortopulmonary window percutaneous closure with an Amplatzer Muscular Septal Occluder device (Abbott),[8] however, the dimension of aortopulmonary window (12 mm) and of device used (16 mm) are the larger ever reported. This kind of device was preferred than Amplatzer Septal Occluder device (Abbott) because characterized both by a major length of central waist as well as by less prominent disks. These features allowed a better adaptation of device to the lesion and a less interference to the aortic and pulmonary vascular wall. The echocardiographic data of last follow-up suggest the safety and effectiveness of transcatheter treatment of this rare defect.

Conclusion

Aortopulmonary window is a rare congenital heart defect. Surgery remains the treatment of choice in adults. However, percutaneous techniques are also feasible. In our case, the Amplatzer Muscular Septal Occluder device (Abbott) allowed to achieve a complete closure without residual shunt and vascular wall injuries during follow-up.

Lead author biography

Dr Mario Giordano is involved in interventional paediatric cardiology and congenital heart disease in adult.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared

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